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Hereditary lower motor neuron disease in a rottweiler puppy: A rare case

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Abstract
A rare case of 2.5 months old female rottweiler puppy was presented with tetraplegia, muscle atrophy and weight loss. The detailed history and neurological examination revealed hind limb weakness (paraparesis) started at 3 weeks of age and progressed to paraplegia after 2 months along with muscle atrophy. The motor neuron disorders are not common in dogs except in young rottweilers, pointers and Britany spaniels, the hereditary lower motor neuron diseases are noticed. The lower motor neuron dysfunction is categorised into accelerated form, intermediate form and chronic form in which accelerated form of dysfunction indicates progressive pelvic limb weakness. Main reason for motor neuron disorders is breed predisposition and inbreeding. There is no treatment available for such cases, recovery is not possible and prognosis is grave.

Keywords: Hereditary lower, motor neuron disease, rottweiler puppy

Introduction
The neurons are said to be messengers of central nervous system which transmits the signals or information to the CNS from periphery (sensory neurons) and from CNS to periphery (motor neurons). The motor neurons are specifically connected to effectors (eg: muscle) [1]. The upper motor neurons originate from the brain and connected to spinal cord segments whereas lower motor neurons transmit the signals of upper motor neurons through the synapse to the effectors. The neuromuscular disorders may originate at any point of this pathway and depending upon the location the diseases are related to UMN or LMN in dogs [1, 2]. The neuromuscular disorders are characterised by progressive limb weakness of forelimbs or hindlimbs (paresis to paralysis), localised or generalised muscle atrophy, ataxia and weight loss [3, 4]. The diseases related to this in young animals are hereditary and breed related involving both the neurons and muscle fibres [5, 6]. The breeds predisposed to NMD's are pointers, rottweilers (LMN dysfunction) and Brittany spaniels (hereditary canine spinal muscular atrophy) [3]. The hereditary LMN disorders are represented as three forms 1) accelerated form - <1 month old puppies show skeletal muscle atrophy and hind limb weakness which progress to tetra paresis within 2-3 months, 2) intermediate form- where the clinical signs start at 4 months of age and slowly progresses to tetra paretic by 2-3 years and 3) chronic form – clinical signs are seen in 1 year old dogs and progression is very slow unlike the above two forms [5, 7]. This case report is about accelerated form of LMN dysfunction in a rottweiler puppy.

Case description
A female rottweiler puppy from a litter of four was presented with tetraplegia along with muscle atrophy and weight loss (Fig 1). At 3 weeks of age this puppy had pelvic limb lameness, ataxia and slowly the muscle mass was getting reduced day by day in spite of having good feed and water intake, urination and defaecation. The hind limb weakness progressed to paraplegia and at 2 months of age the signs got spread rapidly to the forelimbs resulting in tetra-paresis. When this puppy was presented at 2.5 months age all the limbs were paralysed (quadriplegia) with loss of body condition. Based on the history of rapid progression of the disease, young age and breed predisposition the puppy was evaluated for the hereditary lower motor neuron disease. The neurological examination revealed reduced spinal reflexes of both forelimbs and hindlimbs, the atrophy of cervical, thoracic, lumbar and muscles of limbs indicating generalised skeletal muscle atrophy, extensor rigidity of all the limbs, animal was able to lift the head and tail (Fig 2 & 3) with normal perianal reflex. All the above-mentioned symptoms were similar to the ascending form of lower motor neuron dysfunction.

The dog was examined thrice (4, 8, 12 months of age), there was reduction in the growth and body condition (Fig 4) with all the other symptoms remained same from the day of diagnosis. Because it is a hereditary condition there is no particular treatment available and the owner was informed about the poor prognosis.

Fig 1: 2.5 months – tetraplegia and hind limb muscular atrophy

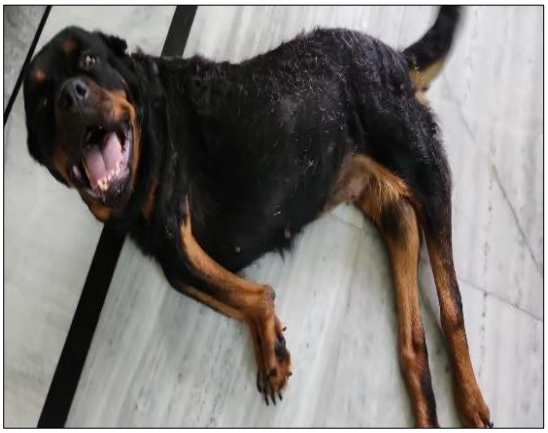

Fig 2: Quadriplegia, generalised muscle atrophy and able to lift the head (8months old)

Fig 3: Quadriplegia, generalised muscle atrophy and able to lift tail with normal intake of water (8 months old)

Fig 4: One year old with reduced body condition

Discussion

Based on the detailed history of rapid progression of disease, clinical signs, neurological examination, body condition score, age and breed related disorder was evaluated and diagnosed as accelerated form of hereditary lower motor neuron disease in 2.5 months old rottweiler puppy. The neuromuscular disorders at young age are mostly degenerative / hereditary in rottweilers and Brittany spaniels [6, 8]. Depending upon the symptoms shown by the dog at initial age and rapid spreading to the other parts of the body, the neuromuscular dysfunction is categorised into accelerated form of disease in which clinical signs are seen in early age of puppies (1 month – 4 months), intermediate form (4-6months to 1 year) and chronic form (>1 year) (7). These clinical signs resemble other neuropathies like Hereditary canine spinal muscular atrophy, the autosomal dominant disease related to motor neurons in Brittany spaniels [3, 4, 5] neuronal abiotrophy in Swedish Lapland dogs [9], the myotubular myopathy (X linked) due to mutation of axon 11 of MTM1 gene in rottweilers puppies with progressive muscle atrophy, tetra paresis and ventroflexion of head, normal proprioception, able to stand for few seconds with short stride gait and cardiomyopathy but all these signs were noticed in 8-9 weeks old [10], although the X linked muscular dystrophy in canines was observed in different breeds like miniature schnauzer, golden retriever, Irish terrier, Alaskan malamute, German short-haired pointer, Belgian Groenendael, Samoyed, Dalmatian, rottweilers, Japanese spitz and corgis etc [7, 8]. Distal myopathy in rottweilers where the typical palmigrade and plantigrade posture due to tarsal and carpal joint hyperflexion along with muscle atrophy and limb weakness noticed in 6-8 weeks old puppies [7, 8]. Idiopathic polyradiculoneuritis and myasthenia gravis are the diseases usually target the motor end plate, in which the former disease is not breed, gender and age related whereas MG is seen in very young and old animals associated with megaoesophagus and mediastinal masses along with neuromuscular symptoms [2]. Because of the similarity in clinical signs, to avoid the difference between neuropathies and myopathies and to describe them briefly, the terms neuromuscular disorders/ motor neuron disorders are used [6].

Conclusion

The neuromuscular disorders in young puppies are very difficult to identify unless those ones related to particular breed like rottweiler where hereditary lower motor neuron disorder is most commonly noticed in litters of inbreeding dogs. The common clinical signs seen in puppies with NMD are quadriplegia, generalised muscle atrophy and weight loss. To determine a particular NMD in these puppies, the complete history of age, gender, breed predisposition, neurological examination, and disease progression are required. Histopathology and especially the molecular level study is required to confirm the type of NMD in animals.

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